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CASE REPORT

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Superior vena caval syndrome caused by the tumor of the left hilum in a patient with unilateral persistent left superior vena cava diagnosed with multislice spiral computed tomography – a case report

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Summary

Background: Unilateral persistent left superior vena cava (PLSVC) is an infrequent finding with incidence of 18-20% among the individuals with PLSVC. The persistence of the left-sided superior vena cava is an effect of disturbances in development of the connection between the precardinal veins (anterior cardinal veins) and formation of the sinus venosus in early stages of embryogenesis.

Case report: The paper presents a case of a 62-year-old patient with a mass lesion of the left hilum, which caused left-sided superior vena caval syndrome in the presence of unilateral PLSVC.

Conclusions: Developmental mechanisms of superior vena caval syndrome are discussed. The evolution of changes related to infiltration and occlusion of PLSVC is shown on the basis of three selected MSCT examinations.

Key words: persistent left superior vena cava • superior vena caval syndrome • lung cancer • multislice computed tomography

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Background

Persistent left superior vena cava (PLSVC) is one of the most common forms of anomalous systemic venous return. It occurs in 0.3-0.5% of the general population, usually as a concomitant vein to the right-sided superior vena cava (RSVC) [1, 2, 3, 4, 5]. Unilateral PLSVC is an infrequent finding with incidence of 18-20% among the individuals with this anomaly [1, 6]. However, Biffi et al. report that approximately 33% of these subjects may not possess RSVC [2]. More frequently, PLSVC is combined with different congenital cardiovascular defects such as atrial and ventricular septal defects, Fallot's tetralogy, transposition of the great vessels or anomalous pulmonary venous return [3, 4, 7]. Its prevalence among these patients reaches 2.8-4.3% [1, 2, 3].

The persistence of the left-sided superior vena cava is an effect of disturbances in development of the connection between the precardinal veins (anterior cardinal veins) and formation of the sinus venosus in early stages of embryogenesis. In normal conditions, after the sinus venosus and right atrium merging at the end of the eighth week of life, the right precardinal vein becomes a privileged one as the shortest way leading the bloodstream to the heart. Therefore, the

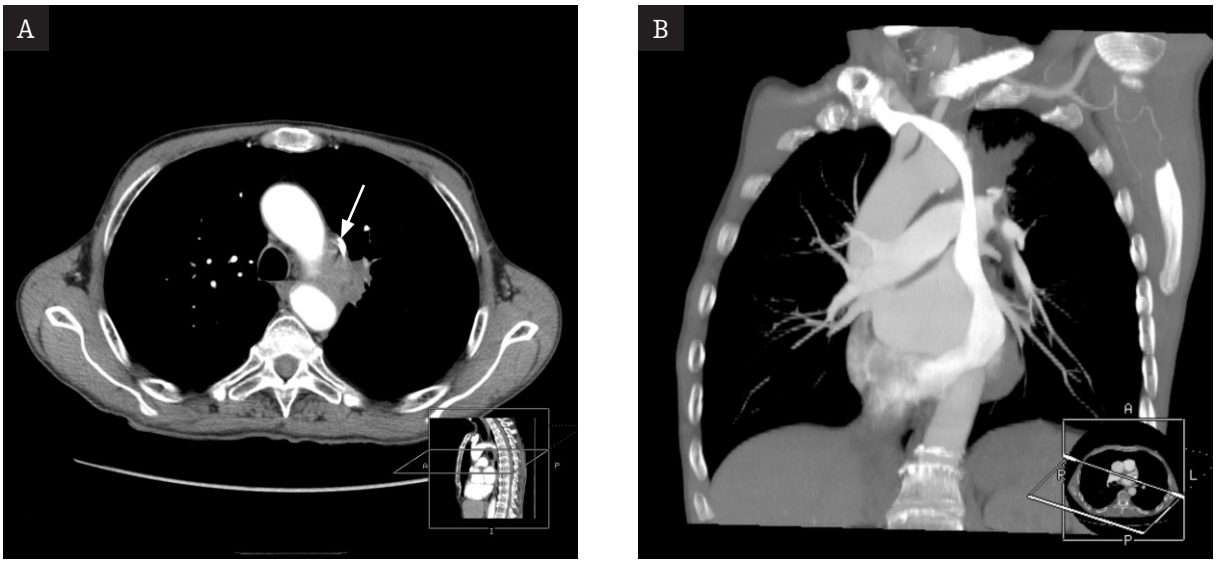


Figure 1. PLSVC infiltrated by the tumor of the left hilum. **A**) Axial scan – the white arrow indicates PLSVC in the neighborhood of the tumor mass. **B**) MIP reconstruction in oblique projection reveals the vein course.

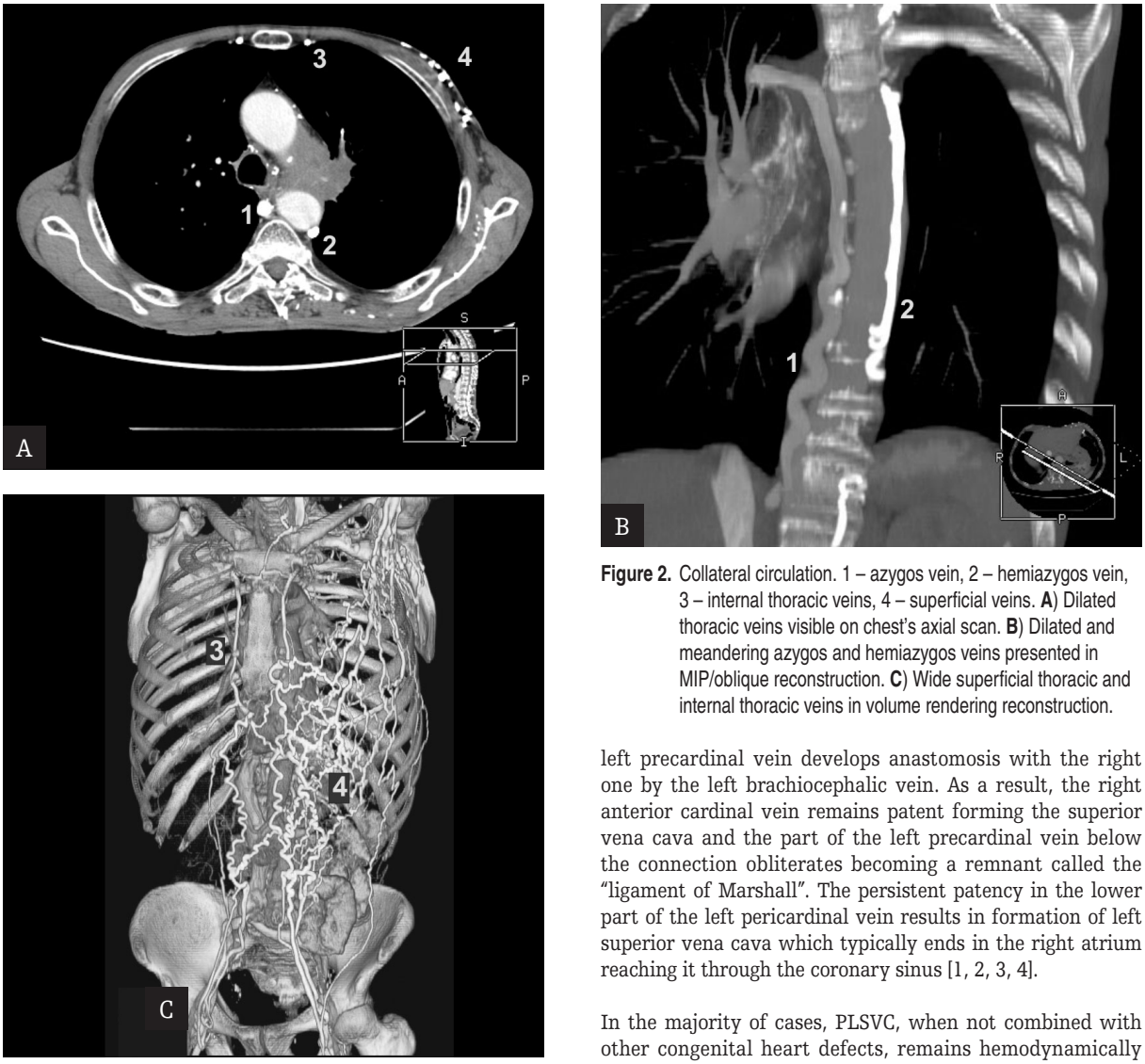
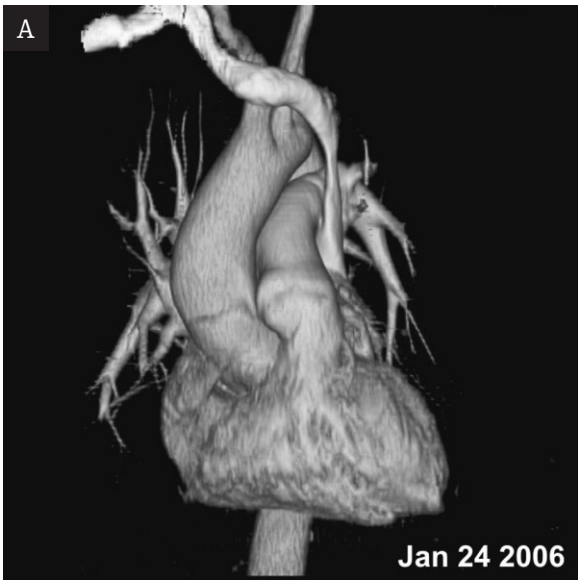
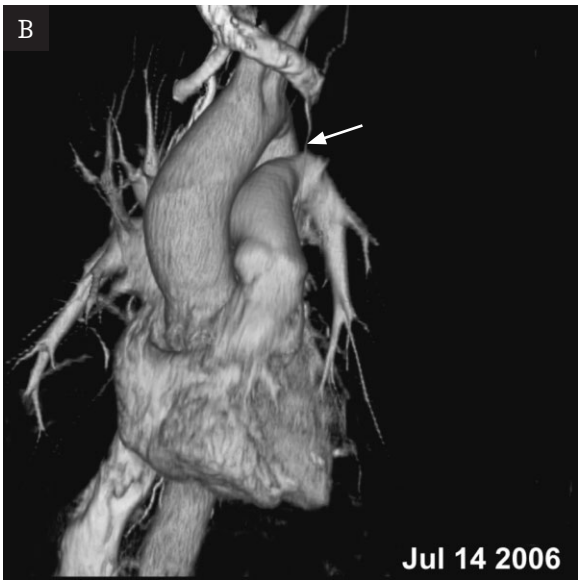


Figure 2. Collateral circulation. 1 – azygos vein, 2 – hemiazygos vein, 3 – internal thoracic veins, 4 – superficial veins. **A**) Dilated thoracic veins visible on chest's axial scan. **B**) Dilated and meandering azygos and hemiazygos veins presented in MIP/oblique reconstruction. **C**) Wide superficial thoracic and internal thoracic veins in volume rendering reconstruction.

left precardinal vein develops anastomosis with the right one by the left brachiocephalic vein. As a result, the right anterior cardinal vein remains patent forming the superior vena cava and the part of the left precardinal vein below the connection obliterates becoming a remnant called the "ligament of Marshall". The persistent patency in the lower part of the left pericardinal vein results in formation of left superior vena cava which typically ends in the right atrium reaching it through the coronary sinus [1, 2, 3, 4].

In the majority of cases, PLSVC, when not combined with other congenital heart defects, remains hemodynamically





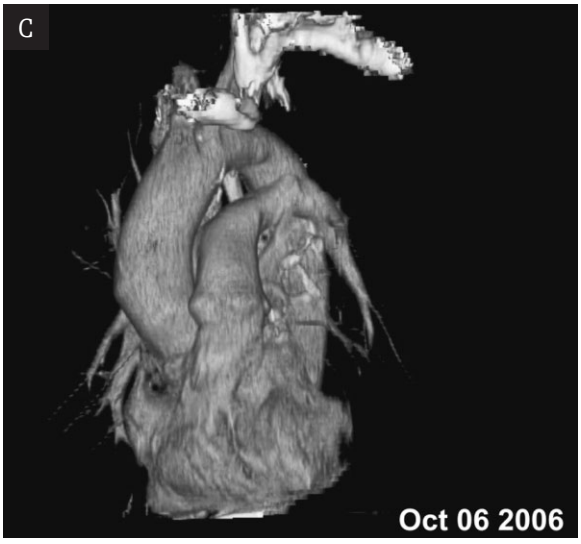


Figure 3. Progressive occlusion of infiltrated PLSVC shown in volume rendering reconstructions. **A**) 24.01.2006 – slight modeling of the vein. **B**) 14.07.2006 – significant narrowing of the vein indicated by white arrow. **C**) 06.10.2006 – complete occlusion of the vein.

asymptomatic [1, 2, 3, 4, 9]. Nevertheless, many authors postulate an association between PLSVC and predisposition to arrhythmias [2, 3, 4, 10]. According to Morgan et al., PLSVC occurs in patients with abnormalities of cardiac rhythm significantly more often than in general population [3]. Clinically, the most important implications of PLSVC are difficulties in cardiac catheterization [1, 2, 4, 5, 6, 8, 9, 11]. Simultaneously, PLSVC is revealed most frequently during this procedure, usually as an incidental finding [4, 11, 12]. The diagnosis of the anomaly is also commonly performed by transthoracic and transesophageal echocardiography, magnetic resonance and computed tomography [13, 14, 15].

Case report

A 62 year – old male, current smoker (20 cigarettes per day, for 40 years) have been hospitalized in order to diagnose a tumor of the left lung, found on chest x-ray. The diagnosis of squamous partially large cell lung carcinoma was obtained by bronchoscopy in April 2005. The patient underwent CT examination of the chest with an 8-row tomograph. The examination was performed in two phase protocol after administering the intravenous bolus of 80ml of non-ionic contrast medium with an automatic syringe at the speed of 4 ml/s. The area of scanning covered the thoracic cavity and the epigastric region. The following parameters were used: collimation – 2.5 mm, tube rotation time – 0.8 s, 120 kV, 350 mA. Multiplanar and 3D reconstructions were acquired using dedicated workstations.

The examination revealed the presence of an extensive tumor localized approximately to the upper pole of the left hilum. In addition, solitary PLSVC reaching the right atrium through the coronary sinus was found. Slight impression and modeling of the PLSVC by the tumor were noticeable [Fig. 1]. The patient was qualified to chemotherapy. Six cycles of chemotherapy were administered according to the Cisplatin and Etoposide scheme. In January 2006, clinical and radiological progression of the disease was observed. As a second-line treatment, Erlotinib was prescribed. Erlotinib is a new registered anticancer molecule, a selective inhibitor of the epidermal growth factor receptor (EGFR) tyrosine kinase [16, 17, 18]. From January to October 2006 six follow-up examinations in six- to twelve-week intervals were performed using a 64-row tomograph. The scanning area included the thoracic, abdominal and pelvic cavities from the apices of the lungs to the lower margins of the ischia (120 ml of contrast medium, 4 ml/s, tube rotation time 0.5 s, 120 kV, automatic mAs adjustment).

CT examination performed in June 2006 presented the sings of infiltration of the PLSVC, causing significant narrowing of the vein. Although the vessel still remained patent, collateral circulation involving azygos/hemiazygos, mediastinal and superficial veins of the left part of thorax and abdomen developed [Fig. 2]. No clinical symptoms of the superior vena caval syndrome (SVCS) were present at that time.

Another CT examination performed in October 2006 revealed complete occlusion of the PLSVC with presence of strong collateral venous circulation through the routes as mentioned above. Clinically, slight SVCS signs, like edema of the face and upper extremities with visible dilation of the veins, cough and shortness of breath were present.

The evolution of changes related to infiltration and occlusion of PLSVC is shown on the basis of three selected examinations [Fig. 3].

Discussion

The anatomic variants of systemic venous return such as PLSVC are not well-known to all of the radiologists and clinicians while in some situations this knowledge may be extremely important. Although in our case lung cancer of the left hilum was diagnosed before it caused occlusion of the PLSVC, the situation when the SVCS is manifested and the only radiographic findings on the x-ray picture are changes of the left hilum is probable. In such a case, the examinations enabling the visualization of the thoracic great vessels should be considered with respect to finding PLSVC. One of the most appropriate diagnostic techniques seems to be multislice spiral computed tomography (MSCT) of the chest [11, 12], all the more since the detection of PLSVC by plain chest radiographs is considered very difficult [15]. MSCT allows either the recognition of anomalies of the systemic venous return or the diagnosis of pathological changes of the lung-hilum, such as tumor or enlargement of the lymph nodes. In addition, this diagnostic technique gives an opportunity to detect coexisting changes such as metastases or collateral blood flow. Moreover, the connections between the systemic veins and the heart, as well as their neighboring structures, should be evaluated precisely in regard to surgical procedure as one of the methods of treatment of SVCS [13, 14]. The application of multiplanar and three-dimensional reconstructions seems to be very useful in planning treatment, especially in the cases of complex cardiovascular malformations where the anatomical topography of the mediastinal structures may be atypical [14].

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